Combined Discrete Subaortic Stenosis, Idiopathic Hypertrophic Subaortic Stenosis and Tetralogy of Fallot

AUTHOR(S):

KONISHI, YUTAKA; TATSUTA, NORIKAZU; MIKI, SHIGEHITO; CHIBA, YUKIO; KAO, CHIN-TZER; HIKASA, YORINORI; NISHIOKA, KENYA; UEDA, TADASHI; WATANABE, TOSHIAKI

CITATION:

KONISHI, YUTAKA ...[et al]. Combined Discrete Subaortic Stenosis, Idiopathic Hypertrophic Subaortic Stenosis and Tetralogy of Fallot. 日本外科宝函 1978, 47(3): 401-410

ISSUE DATE:

1978-05-01

URL:

http://hdl.handle.net/2433/208269

Arch Jap Chir 47 (3), 401~410, Mai, 1978

Combined Discrete Subaortic Stenosis, Idiopathic Hypertrophic Subaortic Stenosis and Tetralogy of Fallot

YUTAKA KONISHI, NORIKAZU TATSUTA, SHIGEHITO MIKI,
YUKIO CHIBA, CHIN-TZER KAO, YORINORI HIKASA,
KENYA NISHIOKA, TADASHI UEDA, and TOSHIAKI WATANABE

The 2nd Department of Surgery, Faculty of Medicine, Kyoto University
The Department of Pediatrics, Faculty of Medicine, Kyoto University
The 3rd Department of Medicine, Faculty of Medicine, Kyoto University
(Received for Publication Feb. 3, 1978)

Summary

The unusual occurrence of tetralogy of Fallot, discrete subaortic stenosis and idiopathic hypertrophic subaortic stenosis in an eight-year-old boy is presented.

A total repair of tetralogy of Fallot was successfully performed at the age of four months. However, at operation, we could not find any sign of aortic stenosis although they were not especially sought.

The simultaneous correction of the discrete subaortic stenosis and idiopathic hypertrophic subaortic stenosis was done at the age of eight years. Unfortunately, the patient could not survive due to low cardiac output syndrome in spite of the use of intra-aortic balloon pumping and left heart bypass.

To our knowledge, the combination of discrete subaortic stenosis, idiopathic hypertrophic subaortic stenosis and tetralogy of Fallot has not been reported. Pathogenesis, diagnosis and surgical treatment of this combination of lesions were discussed.

Case Report

This eight-year-old boy was delivered by Cesarean section at 36 weeks. At the age of one month, he developed cyanosis with crying and then frequent episodes of anoxia. Heart catheterization revealed TOF and surgery was performed at the age of 4 months. Typical findings of TOF were noted at the operation. There were an infracristal ventricular septal defect (VSD) with a diameter of 18 mm and infundibular pulmonary stenosis due to hypertrophy of the parietal and septal bands, but pulmonary valvular stenosis was not significant. Under deep hypothermia with surface cooling and limited cardiopulmonary bypass, the obstructing infundibular muscles and fibrous tissue were cut away and a VSD

Key words Discrete subaortic stenosis, Idiopathic hypertrophic subaortic stenosis, Tetralogy of Fallot, Secondary hypertrophic subaortic stenosis.
Present address : The 2nd Department of Surgery, Faculty of Medicine, Kyoto University, Sakyo-ku, Kyoto 606, Japan.

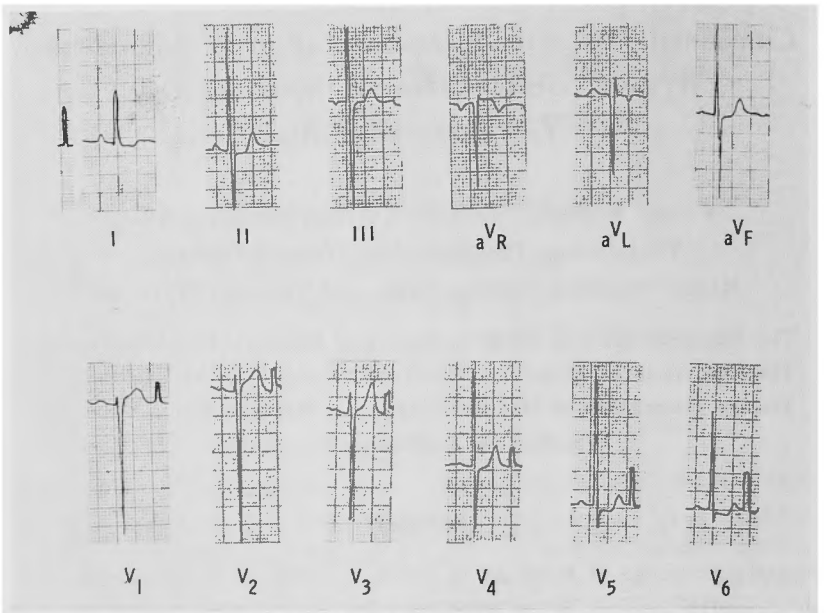

Fig. 1 Electrocardiogram showed left ventricular hypertrophy with ST depression in V_4-V_6.

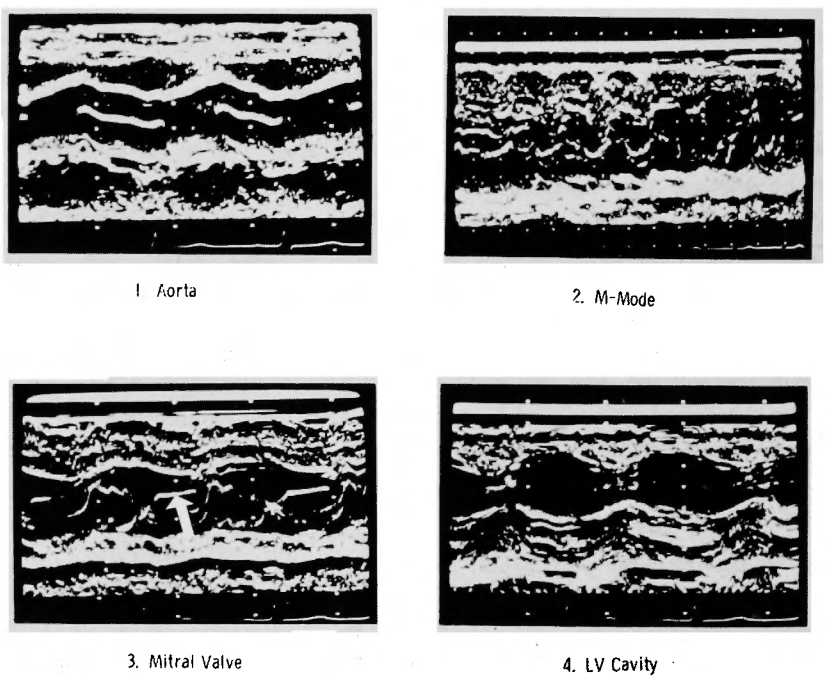

Fig. 2 Echocardiography revealed left ventricular hypertrophy and systolic anterior motion of anterior leaflet of the mitral valve (arrow). The septal wall was much thicker than the posterior wall.

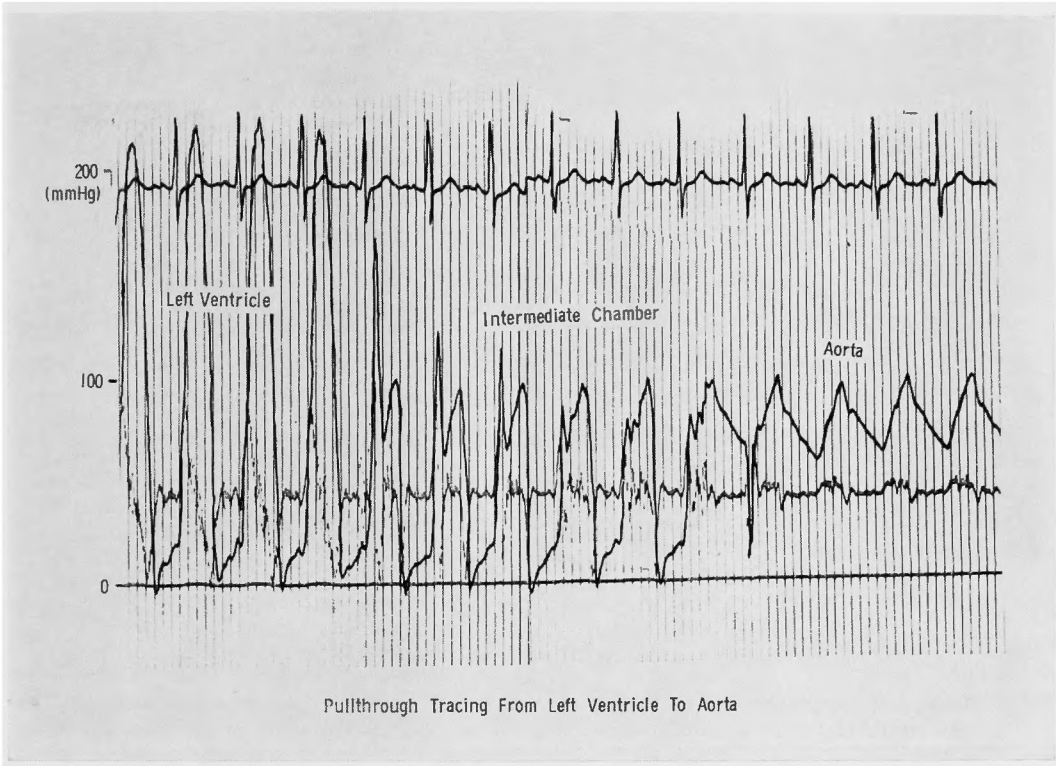

Fig. 3 The pullthrough pressure tracing from the left ventricle to the aorta showed an intermediate chamber.

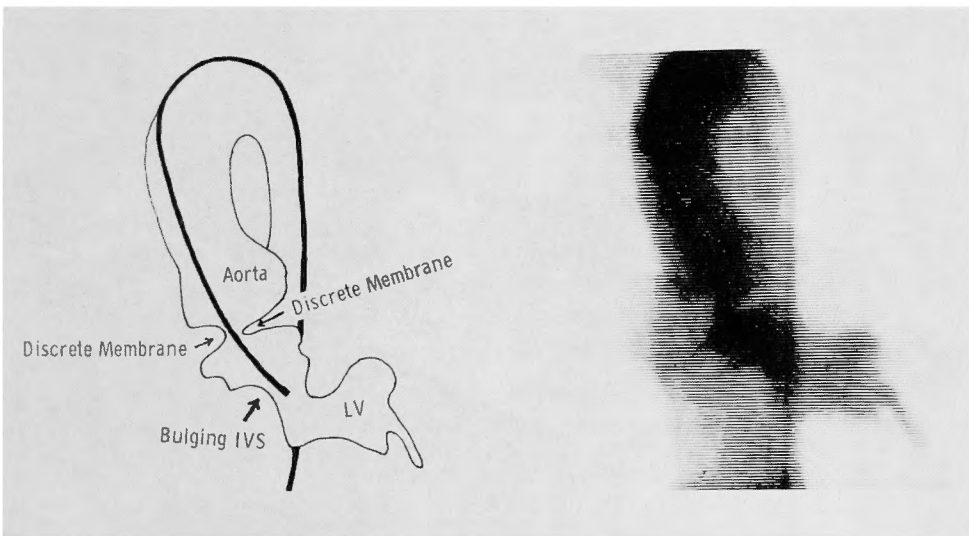

Fig. 4 Left ventriculography revealed discrete membranous obstruction and abnormal bulging of the interventricular septum.

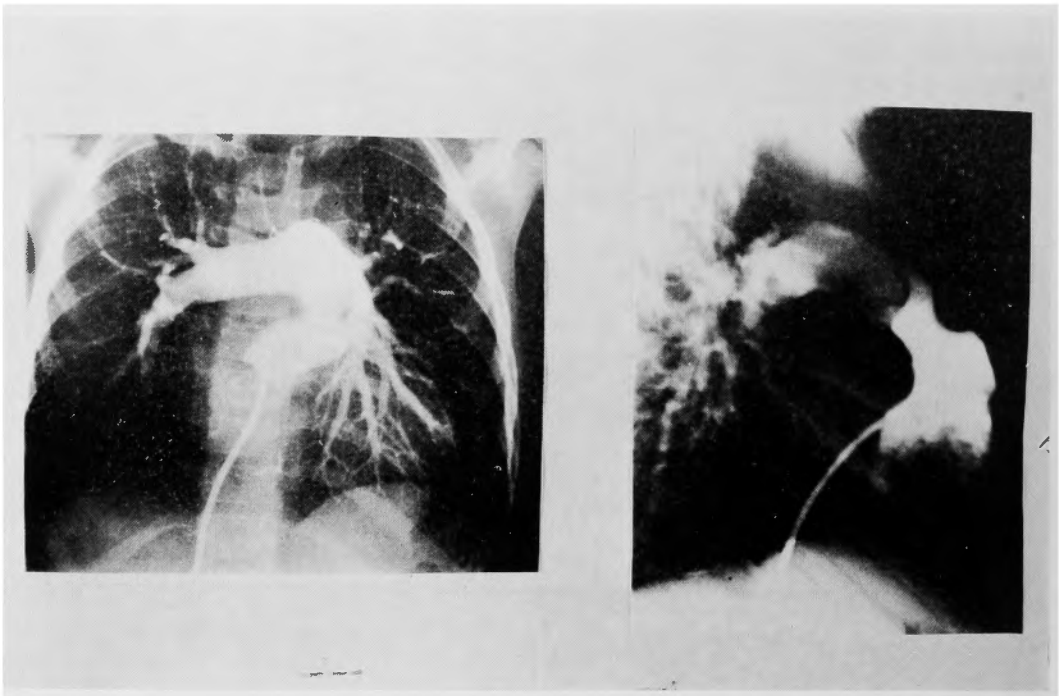

Fig. 5 Right ventriculography demonstrated residual pulmonary valvular and infundibular stenosis. The right ventricular wall was strongly adhered to the sternum. At operation, an aneurysm was found at this area.

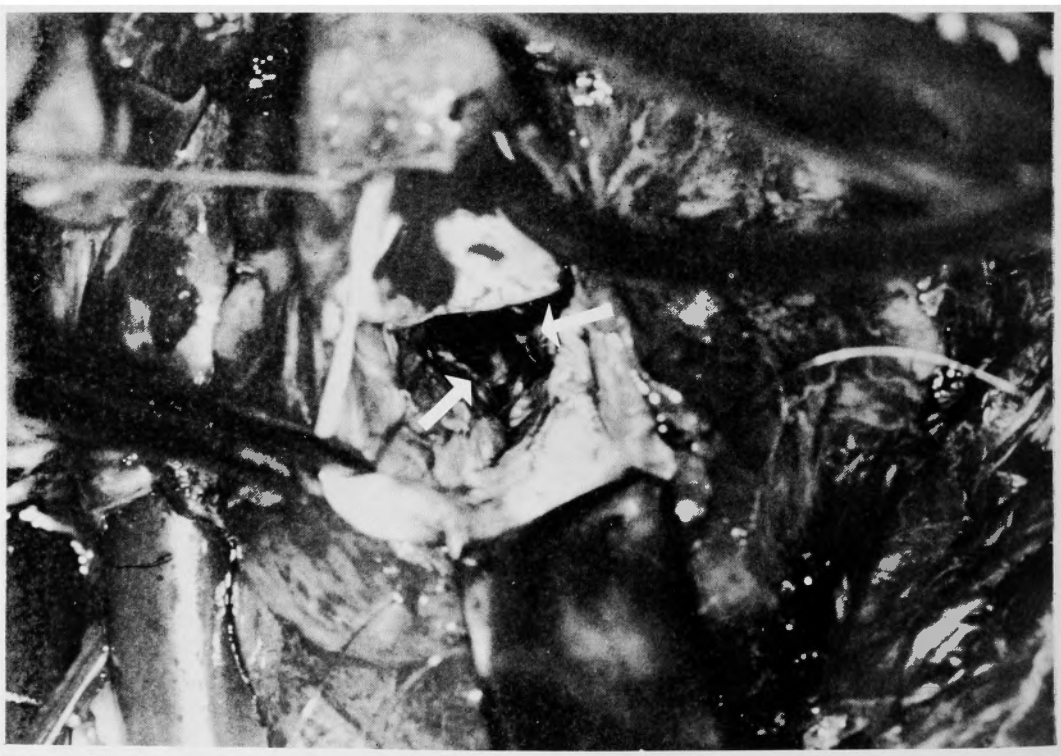

Fig. 6 A discerete membrane (arrows) was found immediately beneath the aortic valve.

Fig. 7 The membrane was excised. Then, the bulging of the interventricular septum was revealed. (arrow).

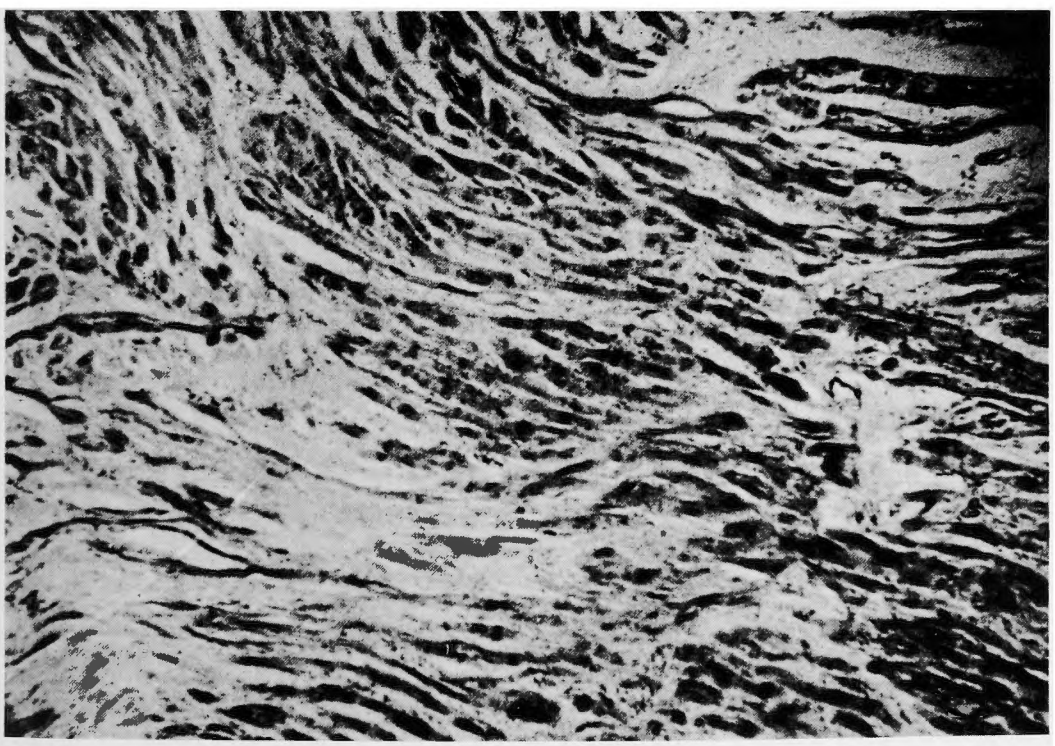

Fig. 8 Hypertrophy and disarray of myocardial fibers were evident. Interstitial fibrosis and the variation of the size of nuclei were also present.

was closed with a pericardial patch. An outflow patch was not required. The postoperative course was uneventful except that he suffered serum hepatitis three months after the operation.

He had been relatively well without cyanosis until eight years of age when he began to have dyspnea on exertion and occasional anterior chest pain. Physical examination revealed a blood pressure of 100/60 and a grade 5/6 harsh systolic murmur at the second right intercostal space, but no diastolic murmurs were audible. A systolic thrill was palpable at the suprasternal notch. The chest X-ray showed cardiomegaly with left ventricular prominence and prominence of the aorta. The electrocardiogram revealed severe left ventricular hypertrophy with ST depression in V_4-V_6 (Fig.1). Echocardiography disclosed left ventricular hypertrophy and systolic anterior motion of the anterior leaflet of the mitral valve. The septal wall was much thicker than the posterior wall (Fig. 2).

At cardiac catheterization, there was a pressure gradient of 104 mmHg between the ascending aorta and the left ventricular cavity. The pullthrough tracing from the left ventricle to the aorta showed the presence of an intermediate chamber (Fig. 3). The subendocardial flow index (SEFI)[23], which was obtained by dividing the area between the aortic pressure and left ventricular pressure during diastole by the integral of the left ventricular pressure during the systolic ejection period, was 0.25. A left ventriculography revealed subvalvular stenosis due to a discrete membrane and the abnormal bulging of the interventricular septum (Fig. 4). On right ventriculography, there was residual pulmonary stenosis of the valvular and infundibular types (Fig. 5). However, the cardiac index was normal and no residual shunt was detected.

Because of these findings, operation was performed. The heart was exposed through a median sternotomy with some difficulty due to adhesions from the previous surgery. An intense thrill was palpable over the aorta positioned anteriorly to the pulmonary artery. There was a ventricular aneurysm with a diameter of 3 cm on the right ventricle where ventriculotomy had been made at the initial surgery. Intracardiac pressure measured at operation revealed a peak systolic pressure gradient of 135 mmHg across the left ventricuar outflow tract.

Prior to the cardiopulmonary bypass, an intra-aortic balloon pumping (IABP) was started to improve subendocardial blood flow preoperatively in an effort to protect the hypertrophied myocardium from anoxia during aortotomy. Moreover, IABP was continuously used during and after the surgery combined with or without cardiopulmonary bypass.

Aortotomy was performed under cardiopulmonary bypass with topical cooling and coronary perfusion. Subaortic discrete membranous obstruction with an orifice of 7 mm in diameter was identified immediately beneath the normal aortic valve (Fig. 6). After the excision of the membrane, we found the hypertrophied interventricular septum protruding so as to obstruct the outflow tract of the left ventricle (Fig. 7). According to the method of MORROW et al.[14]., two vertical parallel incisions 5 mm apart, 5 mm deep and 20 mm long were made in the septum. This tissue was excised. Then, the index finger

could freely pass into the ventricle. The interventricular examination revealed two small VSDs of muscular type near the apex. These VSDs were closed through a left ventriculotomy at the apex. Finally, a right ventricular aneurysm was resected.

Upon the completion of these procedures, an attempt was made to take him off the cardiopulmonary bypass but the patient could not sustain a pressure above 50 mmHg even with the support of IABP. At this time, there was a residual pressure gradient of 35 mmHg between the aorta and the left ventricle.

Assisted circulation was carried on for the next few hours without any improvement of cardiac function and then the patient was placed on left heart bypass. In spite of all efforts, low cardiac output continued with subsequent demise.

Histologic examination of the resected portions of the septum showed hypertrophy and a disorderly array of the myocardial fibers. Interstitial fibrosis and a variation in nuclear size were also present (Fig. 8).

Discussion

The coexistence of fixed left ventricular outflow obstruction and hypertrophic subaortic stenosis may be more frequent than is generally appreciated. More than 30 cases of this combination have been reported[2-6)9-11)16)18-20)].

The clinical diagnosis of hypertrophic subaortic stenosis coexisting with fixed aortic stenosis presents difficulties as the latter may mask the findings of the former. Cardiac catheterization could not always demonstrate both sites of the obstruction and in some cases the correct diagnosis was made at operation or during the postoperative period because of the persistent outflow tract obstruction after the relief of fixed stenosis[2)4)6)11)18)19)]. However, using echocardiography which is diagnostic in IHSS, preoperative diagnosis can now be established[3)5)9)10)16)20)].

It remains speculative whether fixed left ventricular outflow obstruction predisposes to the development of hypertrophic subaortic stenosis or whether the two pathologic states originate independently. Some authors[4)6)18)19)] suggested that the hypertrophy was secondary because the hypertrophic obstruction could resolve with time after the relief of fixed obstruction. On the other hand, BLOCK et al[2)]. reported a case which developed asymmtric hypertrophic stenosis long after the removal of a subaortic membrane. They considered that this probably reflected underlying hypertrophic cardiomyopathy not clinically evident at the time of surgery. BLOOM et al.[3)]. described their two cases as secondary hepertrophy because of findings of symmetric septal hypertrophy and normally arranged muscle fibers in the histology of the resected septum.

In our case, left ventriculography showed an unusual feature of a left ventricular outflow tract due to a discrete membrane and bulging interventricular septum. The pull-through pressure tracing from the left ventricular cavity to the aorta demonstrated an intermediate chamber in the left ventricular outflow tract. In addition, characteristic findings of IHSS were obtained on echocardiography. Our diagnosis was confirmed at

operation. Microscopic examination of the excised septal tissue revealed similar findings seen in IHSS. From the point of view of BLOOM et al.[3], our case might be idiopathic, rather than secondary. Moreover, the membranous obstruction was mild without a significant pressure gradient across the membrane. It was unlikely that the secondary hypertrophy could have developed as a response to pressure overload due to such a mild membranous obstruction.

At the first operation of TOF at the age of 4 months, we could not find any signs of aortic stenosis although they were not especially sought. It was considered, therefore, that aortic stenosis was probably very mild at the initial surgery and subsequently increased in severity as a result of body growth or by an actual decrease in the stenotic area[8].

The combination of discrete subaortic stenosis and other congenital heart disease, such as patent ductus arteriosus[19)21)], VSD[7)13)19)21)], pulmonary stenosis[1)17)], double outlet right ventricle[21)], endocardial cushion defect[7)], corrected transposition of the great arteries[7)] and TOF[15)22)], have been also reported, although the incidence of associated anomalies varied greatly in individual reports. VAN PRAAPH et al.[22)] presented two cases of TOF with severe left ventricular obstruction due to anomalous attachment of the anterior mitral leaflet to the left ventricuar septal surface. A similar case was observed by NAGAO et al.[15)].

However, our case had no abnormal lesions in the mitral apparatus. Thus, it seems likely that, in our case, three cardiac anomalies, namely TOF, discrete subaortic stenosis and IHSS, originated independently although we could not recognized all of them at one time. To our knowledge, this kind of combination has not been published.

With regard to the surgical treatment of coexistent fixed aortic stenosis and IHSS, it is advisable to treat both of these lesions at the same operation[20)]. Surgical removal of obstruction without concomitant septal myotomy and/or myectomy has frequently resulted in an unsatisfactory course due to low cardiac output[2)18)]. However, REIS et al.[19)] suggested that the hypertrophic obstruction, although severe in some patients, did not require treatment and always resolved with time because the hypertrophy was secondary to the fixed obstruction.

Since in our case IHSS was probably more responsible, the simultaneous relief of discrete subaortic stenosis and IHSS was performed. Unfortunatey, the patient did not survive due to low cardiac output in spite of the use of IABP and left heart bypass.

From our experiences in surgery for congenital aortic stenosis[12)], IABP was considered to be useful for protecting and assisting the severely hypertrophied myocardium, especially in the cases with low values for SEFI. The case presented was one of the most severe cases we have ever observed. We should have done surgery at an earlier stage of illness. As pointed by STEWART et all.[20)], delay of operation must result in further left ventricular hypertrophy and deterioration in the left ventricular performance.

Reference

1) Beard EF et al : Combined congenital subaortic stenosis and infundibular subpulmonary stenosis.

Report of a case with successful treatment. Arch Int Med **100**: 647-650, 1957.
2) Block PC et al: Coexistent fixed congenital and idiopathic hypertrophic subaortic stenosis. Am J Card **31**: 523-526, 1973.
3) Bloom KR et al: The association of fixed and dynamic left ventricular outflow obstruction. Am Heart J **89**: 586-590, 1975.
4) Champsaur G et al: Congenital discrete subvalvar aortic stenosis. Surgical experience and long-term follow-up in 20 paediatric patients. Brit Heart J **35**: 443-446, 1973.
5) Cohen IM et al: Combined valvular aortic stenosis, hypertrophic subaortic stenosis and coronary artery disease. Successful surgical correction. J Cardiovas Surg **18**: 241-246, 1977.
6) Davis Het al: Hypertrophic subaortic stenosis as a complication of fixed obstruction to left ventricular outflow. Guy Hosp Rep **119**: 35-45, 1970.
7) Edward JE: Pathology of left ventricular outflow tract obstruction. Circulation **31**: 586-599, 1965.
8) El-Said G et al: Natural hemodynamic history of congenital aortic stenosis in childhood. Am J Card **30**: 6-12, 1972.
9) Harrison EE et al: Coexsting right and left hypertrophic subvalvular stenosis and fixed left ventricular outflow obstruction due to aortic valve stenosis. Am J Card **40**: 133-136, 1977.
10) Johnson AD et al: Combined hypertrophic subaortic stenosis and calcific aortic valvular stenosis. Am J Card **35**: 706-709, 1975.
11) Kirklin JW et al Sugical relief of diffuse subvalvular aortic stenosis. Circulation **24**: 739-742, 1961.
12) Konishi Y et al: The surgical treatment of congenital aortic stenosis. Arch Jap Chir **47**: 94-104, 1978. (Japanese with English abstract)
13) Lauer RM et al: Obstruction of left ventricular outlet in association with ventricular septal defect. Circulation **22**: 110-125, 1960.
14) Morrow AG et al: Oparative treatment in idiopathic hypertrophic subaortic stenosis. Techniques and the results of operative and postoperative clinical and hemodynamic assessment. Circulation **37**: 589-596, 1968.
15) Nagao GI et al: Cardiovascular anomalies associated with tetralogy of Fallot. Am J Card **20**: 206-215, 1967.
16) Nanda NC et al: Echocardiography in the diagnosis of idiopathic hypertrophic subaortic stenosis co-existing with aortic valve disease. Circulation **50**: 752-757, 1974.
17) Neufeld HN et al: Combined congenital subaortic stenosis and infundibular pulmonary stenosis. Brit Heart J **22**: 686-690, 1960.
18) Parker DP et al: Coexistent aortic valvular and functional hypertrophic subaortic stenosis. Clinical, physiologic and angiographic aspects. Am J Card **24**: 307-317, 1969.
19) Reis RL et al: Congenital fixed subvalvular aortic stenosis. Anatomical classification and correlations with operative results. Circulation **43, 44**: 11-18, 1971 (suppl. I).
20) Stewart SS et al: Simulaneous operative correction of aortic valve stenosis and idiopathic subaortic stenosis. Circulation **51, 52**:34-39, 1975 (suppl. I).
21) Takeuchi S et al: Discrete subaortic stenosis associated with intracardiac abnormalities. Successful treatment of two patients. J Thorac Cardiovas Surg **68**: 292-297, 1974.
22) Van Praagh R et al: Tetralogy of Fallot with severe left ventricular outflow tract obstruction due to anomalous attachment of the mitral valve to the ventricular septum. Am J Card **26**: 93-101, 1970.
23) Vincent WR et al: Left ventricular subendocardial ischemia in severe valvar and supravalvar aortic stenosis. A common mechanism. Circulation **49**: 326-333, 1974.

和文抄録

限局性大動脈弁下狭窄症，特発性肥厚性大動脈弁下狭窄症およびファロー四徴症の合併

京都大学医学部外科学教室第2講座
小西 裕，龍田 憲和，三木 成仁
千葉 幸夫，高 欽沢，日笠 頼則
京都大学医学部小児科学教室
西岡 研哉，上田 忠
京都大学医学部内科学教室第3講座
渡辺 俊昭

限局性大動脈弁下狭窄症と肥厚性大動脈弁下狭窄症の合併は従来稀な疾患とされていたが，現在まで30例以上の報告がなされている．しかし上記疾患に加えてフォロー四徴症の合併は今迄報告されていない．

我々は8才の男子の上記合併疾患を経験したので，文献的考察を加えて報告する．

患児は生后4ケ月目にフォロー四徴症の診断で根治術施行．典型的ファロー四徴症の所見をえて，室稜下部心室中隔欠損口の閉鎖，漏斗部狭窄筋の切除を行なった．このときは大動脈狭窄症の存在には気付かず，恐らく極く軽度であったのではないかと想像される．

8才になって運動時の呼吸困難および胸部痛を主訴とし，心臓カテーテル検査を再度施行．左室大動脈圧差 104mmHg を記録し限局性大動脈下狭窄症および特発性肥厚性大動脈弁下狭窄症（IHSS）が疑われ超音波検査でこれを確認した．

手術では膜様狭窄部の切除，Morrow らの方法による肥厚心室中隔の切開および切除を行なった．術后低拍出症候群のため大動脈バルーンパンピングおよび左心バイパスによる補助にも拘らず死亡した．

限局性（固定性）大動脈弁下狭窄と肥厚性大動脈弁下狭窄症の合併は，診断，病因および手術に関して，いくつかの問題が提起されている．すなわち，従来本合併疾患は術中あるいは術后早期に残存圧差より発見されることが多かったが，近年超音波検査の発達により術前診断が可能となった．発生学的には本疾患は2つの狭窄が別々に独立して存在したとする説と，固定性狭窄に続発して二次的に心筋の肥厚性狭窄が生じたと考える説がある．我々の症例では超音波検査の所見，切除心筋の組織像により二次的なものより原発性（IHSS）のものと考えられた．更に限局性膜様狭窄は軽度で，このような軽度狭窄に二次的心筋肥厚が発生するとは考えにくい．

手術に関して，肥厚性狭窄が二次的と考えるものは心筋の切開あるいは切除は必要としないと述べているが，多くの人達は同時に二つの狭窄部の除去を勧めている．

残念ながら，我々の症例は低拍出症候群にて救命出来なかった．早期発見および早期手術の必要性を痛感している．